Retroperitoneal Sarcoma

Fact, Opinion, and Controversy

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KEYWORDS

• Retroperitoneal sarcoma • Soft tissue sarcoma • Surgery • Local recurrence • Radiation therapy • Chemotherapy

KEY POINTS

- Retroperitoneal sarcomas (RPS) are rare cancers whose work-up includes detailed radiologic assessment (CT scan of chest/abdomen/pelvis) and expert pathologic review.
- The overall goal of a primary RPS resection is gross resection of tumor with en bloc removal of closely associated/involved viscera and retroperitoneal musculature.
- Neoadjuvant chemotherapy and/or radiation may be key components of the therapeutic armamentarium in patients with RPS especially in chemosensitive subtypes or borderline resectable tumors.
- The predominant pattern of failure in RPS is local recurrence, which occurs in 25% to 50% of patients at 5 years and 35% to 60% at 10 years.
- Long-term surveillance should be in centers of expertise because the decision making for RPS recurrence is complex and warrants multidisciplinary input and access to clinical trials.

FACT

Soft tissue sarcomas (STS) are malignant neoplasms that arise predominately from mesenchymal tissues including fat, muscle, fibrous tissue, and blood vessels.[1] Overall, 15% of STS arise in the retroperitoneum. Retroperitoneal sarcomas (RPS) are rare

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tumors with an incidence of 0.5 to 1 cases per 100,000.[2] In general, RPS are sporadic cancers; however, there are several hereditary cancer syndromes associated with STS including Li-Fraumeni and neurofibromatosis type 1. Radiation-associated sarcomas are rare and can arise as a late complication of treatment with a median onset of 10 years.[3,4]

Diagnostic Challenges of Retroperitoneal Sarcomas

One of the challenges in the management of patients with RPS is that most present with advanced disease yet are asymptomatic. It is essential in evaluating an undiagnosed retroperitoneal (RP) mass that other tumors are excluded in particular lymphoma, adenocarcinoma, germ cell tumor, and paraganglioma.[5] After performing a through history and physical examination, obtaining tumor markers (ie, LDH, AFP, βHCG) may aid in making a diagnosis. Although endoscopy is not usually necessary, a percutaneous biopsy may be required for a definitive diagnosis. It is also critical that all patients with RP masses have diagnostic abdominal and pelvic computed tomography (CT) scans performed along with a staging CT chest when malignancy is suspected (**Fig. 1**). MRI is used in cases where CT is contraindicated and/or when it may complement CT, whereas the use of ultrasound alone is discouraged. To date, there is limited utility in use of PET scans in patients with RPS; however, they may be useful in staging other RP tumors, such as lymphoma and adenocarcinoma.

In general, RPS grow by direct local extension into adjacent tissues and structures, often pushing them aside and less commonly invade fascial planes, joints, or bone. They are usually large in size (median size, 20 cm) at presentation.[6] There are more

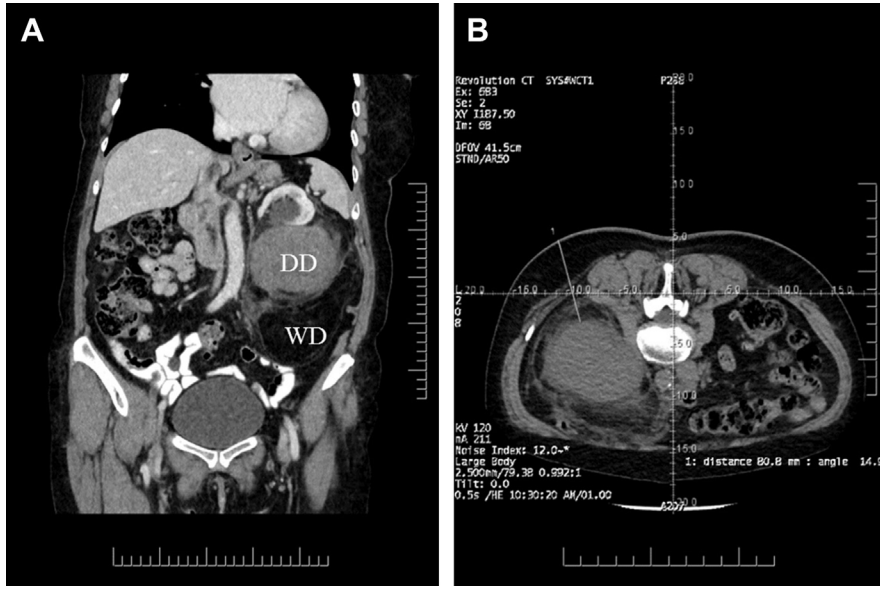

Fig. 1. Establishing the diagnosis of a retroperitoneal sarcoma. Diagnostic CT scan of the abdomen and pelvis demonstrates a retroperitoneal mass with areas suspicious for dedifferentiated (DD) and well-differentiated (WD) liposarcoma (*A*). A diagnostic biopsy was performed using CT-guidance of the high-grade DD area, which confirmed the diagnosis of liposarcoma (*B*).